

The anaesthetic management of the Eisenmenger syndrome

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Summary

The Eisenmenger syndrome is a form of cyanotic congenital heart disease not usually amenable to corrective surgery. It is, however, compatible with leading an active life in early adulthood and due to advances in medical therapy it is not uncommon for patients with this syndrome to live to 30 years or more. Occasionally, therefore, anaesthetists and surgeons will be required to care for these patients when they present for incidental surgery. This article reviews the appropriate measures for the safe operative and perioperative care of patients, based on an understanding of the pathophysiological changes which occur in the Eisenmenger syndrome.

Introduction

In 1897, Dr Victor Eisenmenger (1) described a typical example of what has since become known as Eisenmenger's complex. Necropsy findings were a 2–2.5 cm ventricular septal defect, a large right ventricle, considerable atherosclerosis of the pulmonary artery and its branches, but none in the aorta. There was extensive haemorrhagic infarction of the lungs, secondary to multiple thrombosis. In the Croonian Lecture of 1958, Wood defined Eisenmenger's complex as pulmonary hypertension at systemic level due to a high pulmonary vascular resistance with reversed or bidirectional shunt through a large ventricular septal defect (1.5–3 cm). Wood (2) noted that a similar physiological situation occurred when any communication between the two circulations is complicated by a raised pulmonary vascular resistance sufficient to cause reverse (right-to-left) shunting. The communication may be at aortic level (eg patent ductus arteriosus or aortocopulmonary septal defect), at atrial level (eg total anomalous pulmonary venous return or atrial septal defect) or at ventricular level (eg tricuspid atresia and ventricular septal defect without pulmonary stenosis or a single ventricle. Wood (3) suggested that the expression 'Eisenmenger's syndrome' should be extended to embrace any condition in which there is a communication between pulmonary and systemic circulation that produces pulmonary vascular disease of such severity that right-to-left shunting occurs.

Pathophysiology

In the Eisenmenger syndrome there is decreased cross-sectional area of the pulmonary arteriolar bed with, usually irreversible, pulmonary hypertension. The pulmonary hypertension precludes corrective surgery as the elevated

pulmonary vascular resistance persists or worsens after surgical closure of the defect. This is in contradistinction to patients with obligatory right-to-left shunts such as Fallot's tetralogy when, even in adulthood, corrective surgery may improve the haemodynamic status. Occasionally, in patients with Eisenmenger's syndrome, the increased resistance is not fixed and an attempt to correct the underlying pulmonary-systemic connection may lead to an improved haemodynamic status. The tolazoline test may be used to demonstrate if the pulmonary vascular resistance is fixed or not. (Pulmonary vascular resistance is calculated from the pulmonary blood flow and the pulmonary artery and wedge pressure, before and after the infusion of tolazoline 1 mg kg^{-1} into the pulmonary vein.)

In Eisenmenger's syndrome the pulmonary and systemic vascular resistances are approximately equal and the shunt is balanced. A sudden increase in the right-to-left shunt, associated clinically with an increase in cyanosis, can occur either because of an increase in pulmonary vascular resistance or because of a fall in systemic vascular resistance (4). The factors known to favour the development of pulmonary hypertension are hypercarbia, acidosis, hypoxia, high left atrial pressure and a high pulmonary blood flow (5). Although attempts to decrease the pulmonary vascular resistance in the Eisenmenger syndrome are usually unsuccessful, the systemic vascular resistance remains normal or low and is responsive to physiological and pharmacological influences (6). Any fall in systemic vascular resistance will be adverse, as it increases the right-to-left shunt and thus arterial hypoxaemia. An increase in the systemic vascular resistance will increase the left-to-right shunt and pulmonary blood flow, at the expense of a further increase in pulmonary arterial blood pressure (7).

Young and Mark (8) noted that a number of patients with the Eisenmenger syndrome lived into the third decade and progressively fewer into the fourth, fifth and sixth decades. Patients may therefore present to the anaesthetist when undergoing incidental surgery. Female patients when pregnant may also present if surgical intervention is required.

Anaesthetic considerations

The principle of any anaesthetic technique chosen for a patient with Eisenmenger's syndrome is to avoid a fall in arterial blood pressure by maintaining both cardiac output and systemic vascular resistance (9). Factors reducing cardiac output are: direct myocardial depression or loss of sympathetic drive to the heart, extreme changes in heart rate and a decrease in venous return.

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Heavy premedication is well tolerated by these patients (9) and will enable the dose of induction agent to be kept to a minimum. In addition because of the risk of infective endocarditis, appropriate antibiotics should be administered before and after any surgical procedure.

Meticulous attention is required to avoid the hazard of paradoxical air embolism, by the anaesthetist in relation to infusion lines and by the surgeon in relation to the opening of large veins or venous sinuses with the patient in a position where vascular air entrainment is possible (10).

Induction of anaesthesia presents the time when a fall in systemic vascular resistance and hypotension are likely to occur. Arm-brain circulation time is short, due to the right-to-left shunt, and therefore agents given intravenously will act very quickly. Barbiturates cause hypotension by a combination of reduced cardiac output as well as decreasing the tone of the systemic capacitance vessels, an effect which is dose-dependent (11). Althesin, though it maintains cardiac output, is liable to cause a similar reduction in systemic arterial pressure to barbiturates, due to a reduction in systemic vascular resistance. Thus barbiturates and Althesin must be used with caution, though heavy premedication will enable the minimum doses of these agents to be used. Systemic hypotension, should it occur during induction, is an indication for an α -adrenergic vasopressor, such as methoxamine or phenylephrine. Ketamine has advantages as it does not reduce systemic vascular resistance and has been used to induce anaesthesia in patients with the Eisenmenger syndrome (12). Etomidate and midazolam are other alternatives with minimal cardiovascular effects.

Due to the decreased pulmonary blood flow in the Eisenmenger syndrome, the rate of rise of the arterial concentration of volatile anaesthetics is much decreased (13). Inhalational induction will therefore be slower and higher concentrations of volatile or gaseous anaesthetics may be needed. This will be more marked for the soluble agents such as diethyl ether, trichloroethylene or methoxyflurane. The modern potent volatile anaesthetics such as enflurane and isoflurane are much less soluble in blood and thus this effect is less significant. However, enflurane, isoflurane and halothane all cause a degree of hypotension, due to varying effects on myocardial contractility and systemic vascular resistance, and thus induction or maintenance of anaesthesia with these as sole agents is not advisable. Though now not widely used, agents such as cyclopropane or diethyl ether which have little effect on systemic vascular resistance may be more suitable choices (9). A technique based on intravenous narcotics (opioid anaesthesia) may be the most suitable method of maintenance of anaesthesia. In these circumstances very low concentrations of halothane or enflurane to ensure lack of awareness will not appreciably affect systemic arterial pressure. Isoflurane, although it has little direct myocardial depressant activity, is best avoided, even in low concentration, because of its marked effect in lowering systemic vascular resistance.

Intermittent positive pressure ventilation can decrease pulmonary blood flow and hence increase the right-to-left shunt if excessive transpulmonary pressures are used. However, provided normal tidal volumes are used with minimal inflation pressures, high intrathoracic pressures can be avoided (9). Lumley, Morgan and Sykes (14) recommended minute volumes of 5–8 litres per minute with tidal volumes of 5.5–6 ml/kg body weight to maintain PaCO_2 within normal limits in adults. The most suitable muscle relaxants are those with minimal effects on the cardiovascular system. Thus atracurium or vecuronium are recommended. Tubocurarine, alcuronium and dimethyl-tubocurarine may all to a varying extent cause hypotension and should be avoided. Pancuronium is an alternative though it not infrequently causes tachycardia, an unwanted side effect.

The choice of a general or regional anaesthetic technique may arise, particularly in obstetric or surgical procedures

in the lower abdomen. Although inadvertent hypotension may occur with both techniques, spinal or epidural blockade, associated with inevitable autonomic blockade and peripheral vasodilatation, would seem to be potentially hazardous. In addition, the direct cardiovascular effects of local anaesthetic agents and vasoconstrictors may be unpredictable in their effects. Nevertheless, epidural anaesthesia has been successfully employed for tubal ligation (7) and caesarean section (15). Crawford (16) recommends epidural block as the technique of choice for pain relief in labour, but argues against a regional technique for caesarean section, the extensive autonomic blockade and prospective hypotension being the principal hazard.

On balance, therefore, we are in agreement with other authors (9, 17) that general anaesthesia is preferable for these patients.

Careful attention is required with fluid replacement; loss of extra-cellular fluid is poorly tolerated (5); however, volume overload may increase the right-to-left shunt, or cause right ventricular failure, or both (18). In addition, because of the high packed cell volume, replacement of blood loss should be by colloid or crystalloid solution and not, in the first instance, by blood.

Pregnancy

In a review of pregnant patients with congenital heart disease, Jones and Howitt (4) noted that in patients classified as having the Eisenmenger syndrome, the maternal mortality was 27%, the highest comparable mortality being 4.3% in patients with coarctation of the aorta and 4.2% in those with Fallot's tetralogy. They suggested that the degree of pulmonary hypertension is the most important factor in maternal mortality figures. The two major hazards facing a pregnant patient with the Eisenmenger syndrome are firstly, a fall in systemic blood pressure which would allow the intracardiac shunt to become wholly from right to left, and secondly, thromboembolism which may fatally interfere with an already embarrassed pulmonary circulation (16). Additional risk factors are the increased blood volume, due to pregnancy, and a high haematocrit (10). The greatest risk occurs during delivery and in the immediate post-partum period.

It is generally agreed that caesarean section should be avoided if possible, as blood loss is unpredictable even in the hands of a meticulous operator, and a sudden fall in the circulating blood volume of a patient with Eisenmenger's syndrome can be fatal. Both general anaesthesia with intermittent positive pressure ventilation and regional analgesia with extensive sensory and autonomic block carry high risks (16). Vaginal delivery is considered the method of choice (16). Provided hypotension is avoided epidural block provides the best method of analgesia as the patient can be spared a painful and exhausting labour. If caesarean section is necessary, however, general anaesthesia is probably to be preferred to a regional technique which would require a high level of block (16).

Thromboembolism is a recognised known hazard of pregnancy in patients with Eisenmenger's syndrome as it can result in a sudden increase in pulmonary vascular resistance. Massive pulmonary emboli are uncommon, but a small pulmonary embolus can trigger off widespread pulmonary vasoconstriction (4). The use of anticoagulant therapy throughout pregnancy to prevent pulmonary emboli is not advised, however, because of the danger of inducing life-threatening haemoptysis from engorged pulmonary capillary loops (19). In addition, the disturbed coagulation system following prophylactic anticoagulation would contraindicate epidural analgesia (16) and anticoagulant therapy in the postpartum period may also cause bleeding problems (10). Early ambulation is possibly as important a measure as anticoagulation (10, 18).

Monitoring

Some authors (9) advocate the routine use of ECG monitoring and frequent measurement of blood pressure by non-invasive techniques only, others (7,17) state that intra-arterial monitoring and central venous catheterisation is essential and one group (20) has suggested that a pulmonary artery flow-directed catheter should be inserted in every patient.

The place of invasive monitoring in Eisenmenger's syndrome is controversial and, as with any form of monitoring, the risks and complications must be balanced against the value of the information obtained. These patients have additional risks from invasive monitoring. For example, as they are polycythaemic, intra-arterial catheterisation may be associated with a higher incidence of post-cannulation thrombus formation. Insertion of central venous catheters has the potential risks of infection and paradoxical air embolus. The complications of pulmonary arterial catheterisation are of particular importance in patients with Eisenmenger's syndrome: (a) pulmonary artery rupture, as patients with pulmonary hypertension are more susceptible to this complication, (b) arrhythmias, which may be life-threatening and (c) embolisation, both due to its profound effect on pulmonary vascular resistance and the possibility of systemic embolisation across a congenital cardiac defect which has been reported with fatal consequences (21).

We recommend the use of intra-arterial monitoring in all patients with the Eisenmenger syndrome as hypotension, unless immediately corrected, will allow the intracardiac shunt to become wholly from right to left, with disastrous consequences. A central venous catheter, although not mandatory, should be considered, as an increase in central venous return, in the presence of a high, fixed pulmonary vascular resistance, would increase the right-to-left shunt, whilst a decrease in venous return would decrease pulmonary and systemic blood flow, both conditions resulting in hypoxaemia (7). The usefulness of a pulmonary artery flow-directed catheter will depend on the operative procedure the patient is undergoing and the site of communication between systemic and pulmonary circulations. Patients with an interatrial or aorto-pulmonary communication will benefit as pressure gradients between the systemic and pulmonary circulations can be monitored (15). In patients with an interventricular communication the situation is different because the ability to monitor both right and left intra-ventricular pressures is unavailable (18). In addition, thermodilution cardiac output determinations may be misleading because of the type and varying degree of shunt and cardiac output must be determined by some other means, such as Fick's principle. Thus the risks of a pulmonary artery flow-directed catheter may not outweigh the benefits of additional information. Because of the potential complications of these invasive monitoring devices, they should be removed when no further useful information is provided (21).

Postoperative considerations

Postoperatively, a rapid return to consciousness and avoidance of hypoxia, which would increase the pulmonary vascular resistance, are essential (19). In addition, avoidance of extreme changes in heart rate, oxygen therapy and early mobilisation (18) is recommended.

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